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Aortic arch reconstruction method with ductus-dependent systemic flow preservation in a critical 'bovine trunk' aortic coarctation treatment

Metoda plastyki łuku aorty z zachowaniem przewodowego przepływu systemowego w leczeniu krytycznej koarktacji o morfologii *bovine trunk*

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Abstract

Introduction. Coarctation of the aorta is a congenital heart defect defined as aortic narrowing at the isthmus. It may occur in association with other defects, which requires a discussion regarding the choice of optimal therapy. Critical aortic coarctation with aortic hypoplasia and so-called bovine trunk anatomy is one of the more challenging forms of this defect, characterized by a common origin of the brachiocephalic trunk and the left common carotid artery, which form a common vessel extending as a direct continuation of the ascending aorta. Therapeutic decisions are made depending on the distance between the ascending aorta and the isthmus and descending aorta, taking into account the typical management of hypoplastic aortic arch.

Material and methods. The clinical presentation in the neonatal period includes weak or absent femoral pulse, pallor, apathy, dyspnea and signs of systemic hypoperfusion with progressive lung edema and acidosis. The diagnosis is mostly based on echocardiography while magnetic resonance imaging or computed tomography allow detailed imaging of the collateral vessels. The most important diagnostic issues are the anatomy of the defect, severity of concomitant aortic isthmus narrowing, and its hemodynamic effect. The medical management is directed at maintaining patency of the ductus arteriosus with prostaglandin E1 infusion and an appropriate balance between peripheral systemic and pulmonary resistance. The recommended treatment of choice is an early surgical correction of the aortic arch defect. The aortic arch reconstruction method with ductus-dependent systemic flow preservation implies resection of the coarctation along with the isthmus narrowing and proximal end-to-side anastomosis, with reconstruction of the inflow to the left subclavian artery using a hypoplastic distal segment of the aortic arch. The operation is performed by the left posterolateral thoracotomy without using extracorporeal circulation.

Conclusions. The prerequisite for selecting a neonate for the proposed procedure is a detailed assessment of aortic arch anatomy, with particular attention to the measurement of the distance between the ascending aorta and the area where the ductus arteriosus connects to the descending aorta. The major advantage of this technique is the ability to perform an off-pump procedure via the lateral access, using only healthy native patient tissues for aortic flow restoration.

Key words: congenital heart defects, paediatric cardiac surgery, coarctation of the aorta, aortic arch hypoplasia

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Introduction

Coarctation of the aorta is a congenital heart defect defined as aortic narrowing at the isthmus [1]. It may occur as an isolated defect or in association with other defects. This seemingly simple anomaly is associated with very varied symptoms, their onset and temporal pattern, potential consequences, and risks for the patient [2]. The current debate on coarctation of the aorta, focused on the optimal surgical technique and timing of surgery, has shifted towards complementary, less invasive approaches based on percutaneous balloon angioplasty and stenting, also in younger patients, and endovascular treatment of thoracic aortic aneurysms. Ongoing controversies arise in patients with coarctation of the aorta accompanying other anomalies, mostly complex intracardiac defects, ventricular septal defect, and various forms of aortic arch hypoplasia.

Anatomy of the defect — critical coartation of the aorta with aortic hypoplasia with bovine trunk anatomy

One of the more challenging forms of aortic arch hypoplasia with coarctation is so called bovine trunk, characterized by a common origin of the brachiocephalic trunk and the left common carotid artery, which form a common vessel extending as a direct continuation of the ascending aorta. In this anomaly, the proximal aortic arch is not identifiable as a separate structure. The distal aortic arch arising from the area of bovine trunk is usually an extremely hypoplastic structure that takes a relatively long course leftward, to the area of the left subclavian artery and the aortic isthmus (Figure 1). Therapeutic decisions are made depending on the distance between the ascending aorta and the isthmus and descending aorta, *i.e.* the length of the hypoplastic distal arch which is usually measured by echocardiography, taking into account the typical management of hypoplastic aortic arch.

Pathophysiology and clinical manifestation

Critical coarctation in neonates, including bovine trunk anatomy

Critical neonatal coarctation is an extreme periductal aortic narrowing leading to advanced heart failure in the first month of life. Closure of the ductus arteriosus in a neonate with isthmus narrowing leads to reduced perfusion of the lower part of the body, manifesting primarily with reduced or absent femoral pulses. The child id pale, apathetic, with signs of peripheral hypoperfusion. Dyspnoea develops due to excessive pulmonary flow and impending pulmonary oedema. Chest X-ray shows evidence of pulmonary congestion and enlarged cardiac silhouette, with electrocardiographic (ECG) signs of right ventricular overload. Arterial

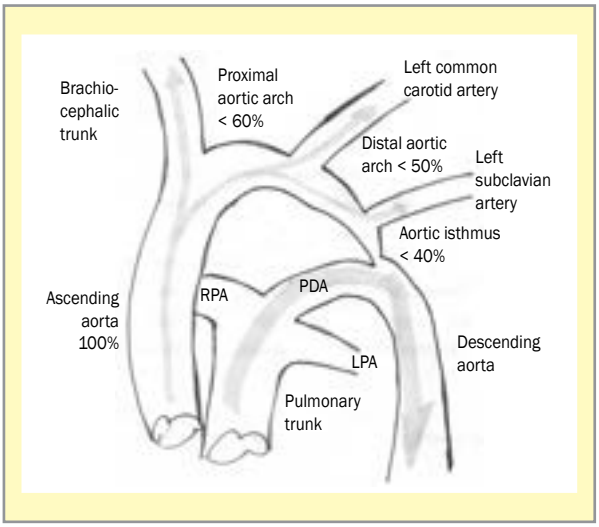

Figure 1. Schematic anatomy of coarctation of the aorta with aortic arch hypoplasia. It is clinically useful to compare the ascending aortic diameter with the arch diameter proximally, distally, and at the isthmus. The diagnosis of aortic arch hypoplasia is based on the reduction of proximal arch diameter below 60%, distal arch diameter below 50%, and the isthmus diameter below 40% of that of the ascending aorta; LPA — left pulmonary artery; PDA — patent ductus arteriosus; RPA — right pulmonary artery

blood gases show progressive metabolic acidosis, initially with respiratory compensation. Arterial oxygen tension (PO_2) is usually normal despite the clinical presentation of pulmonary oedema. Prolonged uncompensated acidosis rapidly leads to secondary multiorgan failure, with renal and hepatic failure, necrotic enterocolitis, and seizures, ultimately resulting in death.

Diagnostic workup and imaging

The most important diagnostic issues are the anatomy of the defect, severity of concomitant aortic isthmus narrowing, and its hemodynamic effect. Collateral circulation is mostly through the intercostal arteries, connecting parasternally with the internal mammary arteries, and posteriorly with the subscapular arteries. Due to resistance generated by severe isthmus stenosis, development of extensive collateral circulation may result in apparently normal femoral arterial pulse, imitating normal aortic flow. In patients with effective collateral circulation, comparative blood pressure measurement, either by a sphyghmomanometer or invasively during cardiac catheterization, may not show a difference between upper and lower limbs or between the aorta proximal and distal to the narrowing. Thus, patient selection based only on blood pressure measurements, without precise delineation of the aortic anatomy, is not sufficient.

Echocardiography

Two-dimensional echocardiography allows excellent imaging in the youngest children, neonates and infants. It is currently the standard modality in neonates with this defect, usually sufficient for proper patient selection for surgical treatment. Typical Doppler echocardiography finding in coarctation of the aorta include absence of typical diastolic flow reduction, replaced by mildly reduced diastolic wave known as the "tail" [3]. An important recommendation to improve safety of neonates with suspected coarctation is to perform the echocardiographic examination repeatedly in the first hours and days of life. Although it may be misleading, echocardiographically confirmed normal flow at the isthmus following spontaneous, often delayed closure of the ductus arteriosus is considered an indicator of patient safety.

Fetal echocardiography

Fetal echocardiography may reveal findings suggesting coarctation of the aorta; in the hands of an experienced prenatal cardiologist, its sensitivity has been estimated at about 30%. Already in early pregnancy, beginning from 16–17 to 20 weeks of gestation, evidence of disproportional vessel size, with predominance of the pulmonary artery, right atrial and ventricular enlargement, and so-called arcade configuration of the aortic arch, indicate a likelihood of coarctation of the aorta in the fetus.

Angiography

Conventional angiography is currently rarely used for the diagnosis of typical coarctation but may be useful in case of echocardiographic uncertainties and in patients with unclear aortic arch anatomy, suspicion of additional defects, and in older patients [4].

Magnetic resonance imaging

The aortic arch and proximal ascending aorta are well imaged by magnetic resonance imaging (MRI). This modality, particularly useful in older children and adult patients (in whom it allow better imaging than any other noninvasive modality), also allows detailed imaging of the collateral circulation, including flow mapping in the aorta and collateral vessels [5].

Computed tomography

Computed tomography (CT) is associated with exposure to large radiation dose, which may be particularly concerning in young children that will require repeated imaging in unclear cases. The major advantage of CT is precise imaging in patients with implanted metal stent, those at a high risk of aneurysm development, and with contraindications to MRI [6, 7].

Drug treatment and preparation for cardiac surgery

The medical management in neonates with diagnosed critical coarctation of the aorta and aortic arch hypoplasia is directed at maintaining patency of the ductus arteriosus and an appropriate balance between peripheral systemic and pulmonary resistance. The patient requires peripheral venous access for prostaglandin E1 infusion at the initial dose of 0.025–0.05 μg/kg/min, usually with dose reduction under echocardiographic monitoring of blood flow in the ductus arteriosus. The main risk associated with prostaglandin E1 therapy are neurological symptoms including seizures and apnoea, often necessitating intubation and ventilation. Ventilation at low FiO_2 and arterial carbon dioxide tension (PCO_2) > 45 mm Hg is recommend until correction of acid-base abnormalities and return of effective diuresis, which may be aided by adding dopamine or dobutamine infusion at a low dose (5 μg/kg/min). In patients who tolerate prostaglandin E1 infusion well and do not require intubation, enteral nutrition is recommended.

The recommended treatment of choice is an early surgical correction of the aortic arch defect.

Surgical treatment

Operative technique — resection of coarctation with isthmus narrowing and proximal end-to-side anastomosis, with restoration of blood flow to the left subclavian artery using the hypoplastic distal aortic arch

The surgery is performed with left posterolateral thoracotomy, with the currently recommended less invasive technique of limited skin incision and muscle protection. With this less extensive chest opening, the serratus anterior and the trapezoid muscles are left outside the main incision line. The chest cavity is opened through the third or sometimes fourth left intercostal space, with somewhat more extensive opening in patients in whom significant collateral circulation is expected. The area of aortic isthmus, distal aortic arch and the left lung hilum is exposed by moving lungs anteriorly. The parietal pleura is opened close to the vessels, allowing aortic mobilization in the area of distal aortic arch, isthmus, ductus arteriosus, descending aorta, and left subclavian artery. The distal aortic arch must be mobilized entirely, along with the initial part of the left common carotid artery. Prominent lymphatic vessels are usually located at the superior aspect of the left subclavian artery, and their damage or incomplete closure using electrocautery may lead to persistent chylothorax in the early postoperative period. A large tributary of the

accessory hemiazygos vein is usually located below, at the aortic isthmus, which must be mobilized and moved sideways during the surgery, or interrupted with closure of both ends in the youngest children. When dissecting vessels, it is helpful to use synthetic atraumatic vascular loops which are temporarily placed under the vessel being freed. Particular attention should be paid to preservation of the vagus nerve that runs at the anterior aspect of the aorta and ductus arteriosus, with identification of the recurrent laryngeal nerve. While the course of the vagus nerve seems consistent, and its trunk is usually a clearly visible and easily identifiable anatomical structure, the course of the recurrent laryngeal nerve may take a number of variants, most commonly looping cephalad under the ductus arteriosus, distal aortic arch, or the left common carotid artery. Preservation of the latter nerve is very important for the patients, particularly the youngest ones, as damage to the recurrent laryngeal nerves results in vocal cord paresis which significantly impairs spontaneous ventilation in the postoperative period. Aortic mobilization requires careful dissecting and freeing all intercostal arteries and collateral vessels within the anterior and posterior walls, and interrupting small arteries supplying the oesophagus, which allows reliable intraoperative aortic clamping and provides additional safety margin in case of an unexpected bleeding during the surgery. Dissecting the posterior aortic wall is associated with a poorly predictable risk of damaging small arteries supplying the spinal cord. The descending aorta is dissected down to the level of several, often dilated intercostal arteries, until it is possible to approximate both aortic ends without undue tension after cutting out the narrowed isthmus and the hypoplastic aortic arch. In older patients, the dilated intercostal vessels may be extremely thin-walled and thus very careful manipulation is advised when freeing them. The last stage is dissection of the ductus arteriosus, with placement of double ligatures for its later closure.

Aortic isthmus narrowing is often accompanied by aortic arch hypoplasia which may significantly limit blood flow to the lower body despite resection of the coarctation with simple end-to-end anastomosis. For this reason, a technique was developed for repairing the isthmus narrowing with concomitant dilation of hypoplastic aortic arch, known as extended end-to-end anastomosis.

In patients with extreme aortic arch hypoplasia, it is necessary to connect the descending aorta with the initial part of the aortic arch using end-to-side anastomosis.

This technique is useful with extreme aortic arch hypoplasia, when aortic isthmus narrowing is accompanied by an anatomic variant known as bovine trunk (Figure 2). In this variant, the ascending aorta extends into a common origin of two vessels, the brachiocephalic trunk and the left common carotid artery, without a discernible proximal aortic arch, considered extremely hypoplastic. Usually, a narrow hypoplastic distal aortic arch extends leftward, giving rise to the left subclavian artery and extending into a critically narrowed aortic isthmus. In this anomaly, the wall of hypoplastic aortic arch is thin and fragile. The descending aorta is a continuation of a very wide ductus arteriosus, which provides effective outflow from the pulmonary artery while the patient receives prostaglandin therapy, sometimes with clearly felt peripheral pulses. After chest opening with typical lateral exposure, the hypoplastic distal aortic arch is usually completed covered by a wide ductus arteriosus, which may imitate normal aortic anatomy.

A repair of this type of anomaly in a neonate is possible using a typical lateral access. Initially, wide dissection is performed to expose all aortic branches supplying the brain, the aortic isthmus area with the ductus arteriosus, and the descending aorta with intercostal arteries down to a very low level. During the first stage, a DeBakey clamp is placed at the aortic isthmus above the ductus arteriosus, and an additional C-clamp is placed on the distal aortic arch with the left subclavian artery (Figure 3). After the isthmus is interrupted above the clamp, the opening in

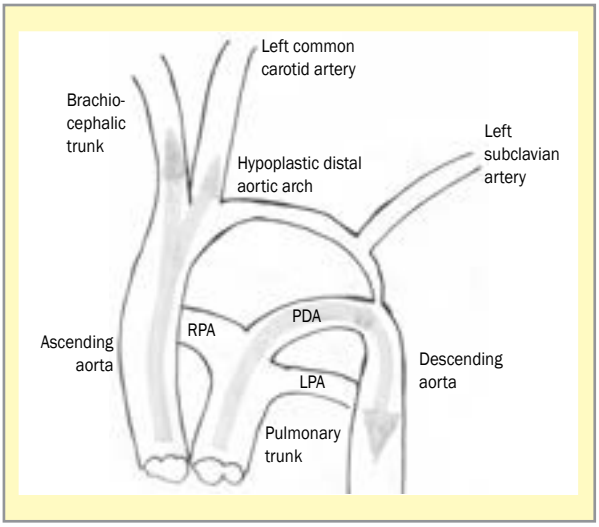

Figure 2. Aortic arch hypoplasia with bovine trunk coarctation. The normal ascending aorta extends into a common origin of two vessels, the brachiocephalic trunk and the left common carotid artery, without a discernible typical proximal aortic arch. The initial part of the aortic arch is thus considered undeveloped (extreme hypoplasia), while the distal arch, connecting to the left subclavian artery and the aortic isthmus, is long, hypoplastic and usually thin-walled. The left subclavian artery originates at the connection of the aortic arch with the critically narrowed isthmus but may be often located more caudally, at the coarctation or even lower, originating from the descending aorta. In this defect, the descending aorta is a continuation of a very wide ductus arteriosus; LPA — left pulmonary artery; PDA — patent ductus arteriosus; RPA — right pulmonary artery

the aortic arch is closed by a suture, and the C-clamp is released. With continuous prostaglandin infusion and preserved patency of the ductus arteriosus, the hemodynamic status remains unchanged at this stage, with the brain and both upper limbs supplied directly by the heart, and the lower body perfused via the ductus arteriosus. Later, after the prostaglandin infusion is stopped and the ductus arteriosus is ligated, the descending aorta is clamped and a wide resection of the narrowed isthmus with periductal tissue is performed. Under brachiocephalic trunk blood flow monitoring (by invasive blood pressure measurement in the right brachial artery), the area of common origin of the common carotid artery from the ascending aorta is closed laterally at the border of the brachiocephalic trunk and the ascending aorta, along with two peripheral vessels supplying the brain. Following lateral opening of the proximal aortic arch at the origin of the brachiocephalic trunk, a circumferential end-to-side anastomosis is performed within the clamps after the descending aorta is pulled up (Figure 4A, B). An additional difficulty may arise if the subclavian artery originates at the narrowed isthmus or even lower, from the ductal tissue. In this setting, an independent transplantation of the left subclavian artery to the left common carotid artery seems warranted with an end-to-side anastomosis. The procedure is concluded by controlling hemostasis within the extensive dissected area (Figure 5).

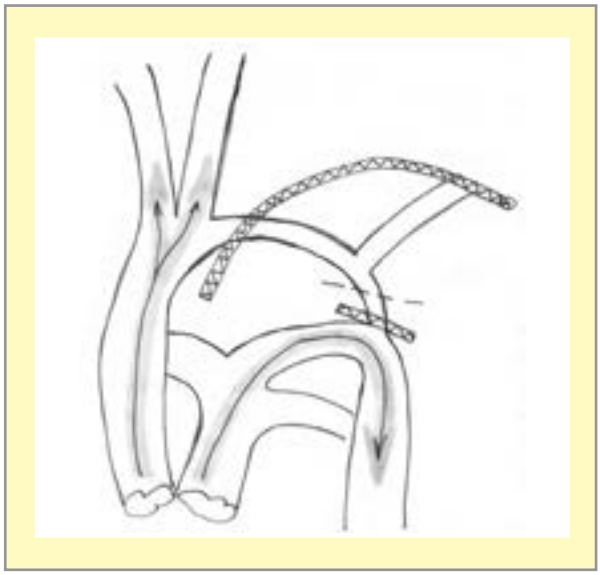

Figure 3. The technique of resecting coarctation with the aortic isthmus and creating a proximal end-to-side anastomosis, with flow restoration to the left subclavian artery using the hypoplastic distal aortic arch. During the initial stage, after wide dissection of all vessels and clamping the hypoplastic aortic arch and the isthmus, the isthmus is interrupted, with its proximal end closure by a continuous suture. After the proximal clamping is released, with continuous patency of the ductus arteriosus during prostaglandin E1 infusion, the hemodynamic status and cerebral and systemic perfusion remain unchanged

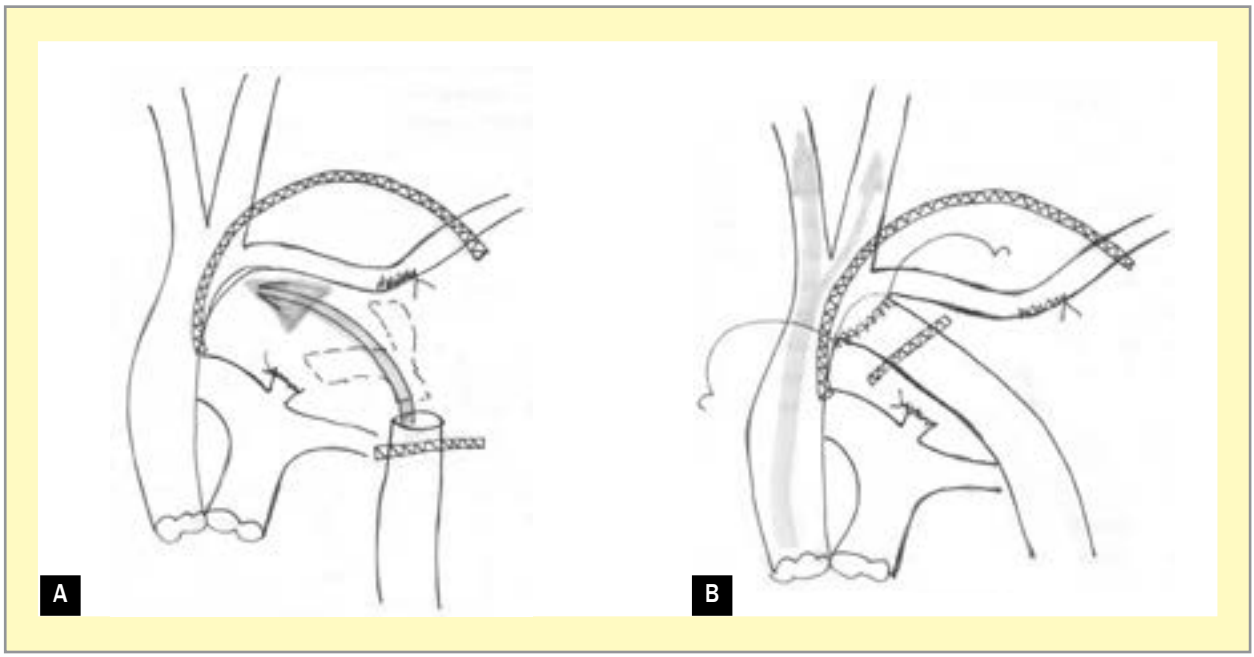

Figure 4A. The next procedural step after clamping is relocated, with moving the proximal C-clamp towards the ascending aorta and tangential brachiocephalic trunk clamping to provide adequate cerebral flow (monitored by invasive blood pressure measurement in the right brachial artery). Aortic wall is opened inferiorly at the distal ascending aorta and the initial part of the hypoplastic aortic arch. Prostaglandin E1 infusion is stopped, and a wide resection of the isthmus area with periductal tissue is performed after ligating the ductus arteriosus and clamping the descending aorta; **B.** An end-to-side anastomosis is performed using a monofilament continuous suture after the clamps are approximated and the descending aorta is pulled upward

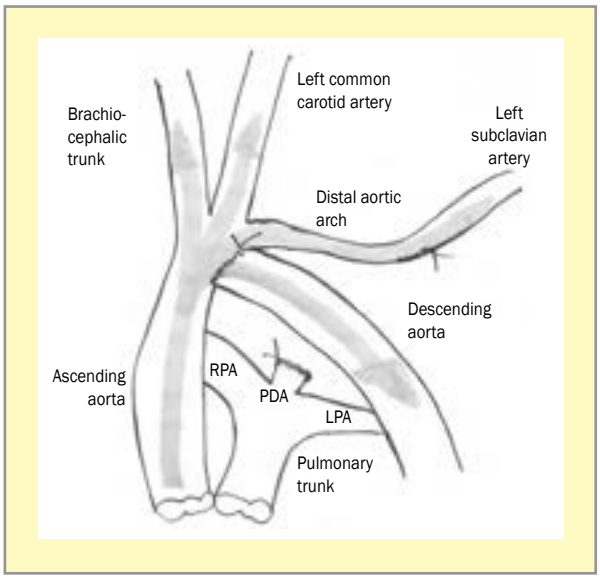

Figure 5. Aortic anatomy after end-to-side anastomosis: the distal hypoplastic aortic arch serves as the initial part of the left subclavian artery, and aortic continuity is restored by proximal anastomosis with the descending aorta; LPA — left pulmonary artery; PDA — patent ductus arteriosus; RPA — right pulmonary artery

The prerequisite for selecting a neonate for the proposed procedure is a detailed assessment of aortic arch anatomy, with particular attention to the measurement of the distance between the ascending aorta and the area where the ductus arteriosus connects to the descending aorta. Optimally, the diagnostic echocardiographic study should be performed by a cardiologist experienced in the preoperative evaluation and patient selection, in cooperation with the cardiac surgical operator. Following end-to-side anastomosis, the inflow to the descending aorta bypasses the hypoplastic aortic arch which only serves to provide aortic continuity and inflow to the left subclavian artery. The major advantage of this technique is the ability to perform an off-pump procedure via the lateral access, using only healthy native patient tissues for aortic flow restoration.

Conflict of interests

The authors declare no conflict of interests.

Streszczenie

Wstęp. Koarktacja aorty jest wrodzoną wadą serca definiowaną jako zwężenie aorty w okolicy cieśni. Wada ta może współwystępować z innymi anomaliami, stanowiąc przedmiot dyskusji nad wyborem optymalnej terapii. Krytyczna koarktacja aorty z hipoplazją aorty o anatomii tak zwanej sylwetki byka (ang. *bovine trunk*) to jedna z trudniejszych postaci wady, w której pień ramienno-głowowy i lewa tętnica szyjna odchodzą wspólnie, początkowo jak wspólne naczynie w kontynuacji aorty wstępującej. Zależnie od odległości aorty wstępującej od cieśni i aorty zstępującej podejmowane są decyzje o sposobie leczenia, z uwzględnieniem postępowania typowego dla hipoplastycznego łuku aorty.

Materiał i metody. Objawy kliniczne pacjentów w okresie noworodkowym obejmują osłabienie lub całkowity brak tętna udowego, bladość, apatię, duszność oraz objawy hipoperfuzji obwodowej z postępującym obrzękiem płuc i kwasicą. W diagnostyce stosuje się przede wszystkim echokardiografię, natomiast rezonans magnetyczny i tomografia komputerowa pozwalają na szczegółowe obrazowanie układu naczyń krążenia obocznego. Najważniejszymi zagadnieniami diagnostycznymi są anatomia wady, towarzyszące jej nasilenie zwężenia cieśni aorty i jego efekt hemodynamiczny. Postępowanie zachowawcze jest ukierunkowane na utrzymanie drożności przewodu tętniczego poprzez infuzje prostaglandyny E1 oraz balansu między oporami obwodowego przepływu systemowego i płucnego. Rekomendowanym leczeniem z wyboru pozostaje wczesna kardiochirurgiczna korekcja wady łuku aorty. Metoda plastyki łuku aorty z zachowaniem przewodowego przepływu systemowego zakłada resekcję koarktacji wraz ze zwężeniem cieśni i proksymalnego zespolenia metodą koniec-do-boku, z odtworzeniem dopływu do lewej tętnicy podobojczykowej z wykorzystaniem hipoplastycznego dystalnego odcinka łuku aorty. Operację wykonuje się z dostępu przez lewostronną tylno-boczną torakotomię, bez zastosowania krążenia pozaustrojowego.

Wnioski. Warunkiem kwalifikacji noworodka do proponowanej procedury jest szczegółowa ocena anatomii wad łuku aorty, ze zwróceniem szczególnej uwagi na pomiar odległości między aortą wstępującą i okolicą połączenia przewodu tętniczego z aortą zstępującą. Podstawowa zaleta tej techniki to możliwość jej wykonania bez zastosowania krążenia pozaustrojowego, z dostępu bocznego, w granicach i z wykorzystaniem zdrowych, własnych tkanek pacjenta.

Słowa kluczowe: wrodzone wady serca, kardiochirurgia dziecięca, koarktacja aorty, hipoplazja łuku aorty

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